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1: Am J Pathol. 2006 Sep;169(3):806-14.

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Odontogenic keratocysts arise from quiescent epithelial rests and are associated with deregulated hedgehog signaling in mice and humans.

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Odontogenic keratocysts in humans are aggressive, noninflammatory jaw cysts that may harbor PTCH1 mutations, leading to constitutive activity of the embryonic Hedgehog (Hh) signaling pathway. We show here that epithelial expression of the Hh transcriptional effector Gli2 is sufficient for highly penetrant keratocyst development in transgenic mice. Mouse and human keratocysts expressed similar markers, leading to tooth misalignment, bone remodeling, and craniofacial abnormalities. We detected Hh target gene expression in epithelial cells lining keratocysts from both species, implicating deregulated Hh signaling in their development. Most mouse keratocysts arose from rests of Malassez--quiescent, residual embryonic epithelial cells that remain embedded in the periodontal ligament surrounding mature teeth. In Gli2-expressing mice, these rests were stimulated to proliferate, stratify, and form a differentiated squamous epithelium. The frequent development of keratocysts in Gli2-expressing mice supports the idea that GLI transcription factor activity mediates pathological responses to deregulated Hh signaling in humans. Moreover, Gli2-mediated reactivation of quiescent epithelial rests to form keratocysts indicates that these cells retain the capacity to function as progenitor cells on activation by an appropriate developmental signal.

PMID: 16936257 [PubMed - indexed for MEDLINE]

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